Cystic Fibrosis strength in numbers

UK CF Registry highlights 2018

2018 Cohort
9,847 Annual Reviews
10,509 Number of active patients

Active patients are people who are currently alive and have submitted an annual review in the last three years.

Life statistics
65 women with CF had babies in 2018
45 men with CF became fathers in 2018
64% people aged 16 years and over were in work or study

Age and sex distribution of the UK CF population in 2008 vs 2018

2008
N=6082
Median age: 18

2018
N=9847
Median age: 20

Overall (%)
Male (%) Female (%)

Note the different demographic distribution across the years, namely higher proportion in older age groups in 2018.
FEV₁% predicted

FEV₁% predicted in 2008 vs 2018

10 year olds
Median FEV₁% predicted

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<thead>
<tr>
<th></th>
<th>2008</th>
<th>2018</th>
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<tbody>
<tr>
<td>Median FEV₁% predicted</td>
<td>86%</td>
<td>91%</td>
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18 year olds
Median FEV₁% predicted

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<th>2008</th>
<th>2018</th>
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<tr>
<td>Median FEV₁% predicted</td>
<td>72%</td>
<td>74%</td>
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30 year olds
Median FEV₁% predicted

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<th>2008</th>
<th>2018</th>
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<tr>
<td>Median FEV₁% predicted</td>
<td>60%</td>
<td>64%</td>
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FEV₁% is reported for people who have not had a lung transplant.

Transplant

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<tr>
<th></th>
<th>2008</th>
<th>2018</th>
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<tbody>
<tr>
<td>Evaluated</td>
<td>126</td>
<td>247</td>
</tr>
<tr>
<td>Accepted</td>
<td>55</td>
<td>104</td>
</tr>
<tr>
<td>Double lung transplant</td>
<td>16</td>
<td>58</td>
</tr>
<tr>
<td>Other transplant</td>
<td>8</td>
<td>7</td>
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Nutrition

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<th>2008</th>
<th>2018</th>
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<tr>
<td>Median BMI Percentile Age 2-17</td>
<td>50</td>
<td>52</td>
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Adults meeting BMI target

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<tr>
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<th>2008</th>
<th>2018</th>
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<tr>
<td>41%  Adults meeting BMI target 2008</td>
<td>48%  Adults meeting BMI target 2018</td>
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The target BMI is 23kg/m² for males and 22kg/m² for females.
Changes in infection rates

Chronic *Staphylococcus aureus* | Chronic *Pseudomonas aeruginosa* | *B. cepacia complex*
---|---|---
15% | 39% | 3%
16% | 27% | 4%

The Registry diagnosis of chronic infection requires three positive sputum samples in the past 12 months.

Lung infections in 2018

Proportion of patients (%) vs Age (years)
The proportion of patients receiving at least one home or hospital IV course in 2018.

Supplemental feeding

- Any
- Gastronomy Tube
- Nasogastric Tube
- Other

Complications

- ABPA
- CFRD*
- Liver disease
- Osteopenia

Respiratory support in 2018

Physiotherapy in 2018

*On CFRD treatment.
Genotype distribution by devolved nation

Mutation frequency

Proportion of people with CF in the UK who carry at least one of these most common five mutations.

99.1% of people have at least one mutation recorded.
A vital function of the UK CF Registry is to make pseudonymised, anonymised and aggregated data available to researchers from recognised institutions, for the benefit of people with cystic fibrosis.

Requests for access to Registry data are evaluated by the independent UK CF Registry Research Committee, which ensures that the requested data are available and the research team are appropriately configured to receive the data in the format requested. The data request form, along with the Data Sharing Policy, and a list of previous data requests can be accessed via the Cystic Fibrosis Trust website.

**UK CF Registry data request service**

**Quality of data**

- **100%** UK CF centre participation
- **99%** Participation of people with CF
- **94%** Active patients with Annual Review
- **99%** Key variables complete
- **98%** Key variables accurate

**Request UK CF Registry data: www.cysticfibrosis.org.uk/registry**

**Working with industry to make medicines safer**

Pharmacovigilance, or ‘drug-safety’, studies monitor the long-term safety of drugs in everyday use, once they have completed clinical trials and been given a license. There are key advantages to cystic fibrosis pharmacovigilance studies, which usually span a five-year period, being run within the UK CF Registry.

**Quality of the study**

The UK CF Registry is a powerful tool for the monitoring of long-term outcomes in a large cohort of people. It allows access to a comparator cohort that are naïve to the study drug, which may not be possible in other models of drug-safety studies. The scope of the information recorded by the UK CF Registry enables detailed comparisons of patient cohorts receiving different medications, with the ability to disentangle drug safety signals from the numerous other aspects of cystic fibrosis.

**Independence**

The principal investigator for each study is an NHS CF physician who provides independent clinical expertise. The UK CF Registry statisticians provide input to the study design, conduct the analysis and contribute to reporting and interpretation of the results. Summary data only (no patient-level data relating to individuals) are provided to the pharmaceutical company, and results and their interpretation are agreed by the experts independent of the industry collaborators.

**References**
